UNILATERAL PARALYTIC CHOREA, WITH A REPORT OF A CASE.[1]

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Of all nervous disorders chorea is probably the most easily diagnosed, for the reason that the symptoms are so striking and characteristic. This statement, however, applies only to the ordinary form of the disease, with the irregular muscular twitchings, the constant unrest and nervousness that are always found in a well-developed case. In the majority of instances the symptoms are so pronounced as to enable the teacher or the mother of the little patient to diagnose St. Vitus' dance even before the physician makes the diagnosis of chorea.

It is quite different, however, in the cases of chorea with ill-defined symptoms when the disease is just developing, and in the cases in which paralytic symptoms predominate over the irritative ones, viz., the jactitations and twitchings. The diagnosis is most difficult and almost impossible when the paralytic symptoms confine themselves to one side of the body and are unaccompanied by the spontaneous and inco-ordinate movements so commonly seen in chorea.

Fortunately these cases are rare, and the diagnosis can be made either by exclusion or by attention to some points ordinarily found in most cases of chorea but disregarded. It is with a view of emphasizing these that I report a rather rare case of unilateral paralytic chorea with a peculiar etiology.

As far back as 1850 the great Todd[2] recognized paralytic chorea, which he describes as follows: "In a large proportion of cases of chorea the choreic movements occur more on one side than on the other, and sometimes they will be altogether confined to one side—the child being hemiplegically affected. In such a case, as happens when the choreic movements have wholly or partially subsided, the limbs that were previously twitching now become paralyzed. The case now resembles in every detail that of ordinary hemiplegia from cerebral causes. But the following points of difference can usually be observed:

"1. The face is often not affected, or slightly so.

"2. There will not be any paralysis of the tongue, but more or less of the peculiar mode of protrusion which characterizes chorea.

"3. The paralyzed limbs will exhibit, even in a very slight degree, the choreic movements."

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[1] Read at a meeting of the Chicago Neurological Society, March, 1906.

[2] Clinical Lectures on Paralysis, 1850, p. 313.

Gowers[1] reports five cases of paralytic chorea in which one arm was usually more affected than the opposite one, and in which seven to fourteen years was the age of predilection. He says: "Whenever a child of this age suffers from gradual loss of power in the arm and presents no weakness in the face, tongue, or leg, the disease as far as I have seen is always chorea. If the nature of the case be suspected, confirmatory evidence—slight occasional spasm—if looked for, will commonly be at some time detected, either in the weak arm or in the other. The course of these cases is tedious, and as the choreic movements may increase as the weakness passes off, the patients often seem, to the friends, to be getting worse when really getting better. I have never seen this form pass on into severe general chorea."

Osler[2] says: "Muscular weakness is a prominent symptom of chorea and may result in a condition of paresis or even paralysis. It may be hemiplegic, paraplegic, or—more frequently—monoplegic. Very rarely does the loss of power reach the grade of complete paralysis, but the child may be unable to lift the arm to the head, a shoulder may droop, the grasp may be feeble, or there is wrist-drop. It is important to remember that there may be extreme paresis, with slight inco-ordination and few spontaneous movements. Weakness of the arm is shown by enfeebled grip, of the leg by a dragging or by a decided limp. The paralytic symptoms may precede or come on in the course of the attack, more rarely follow it."

The cited descriptions of paralytic chorea are certainly clear and should help one to make a diagnosis in every case. However, if the disease appears after a trauma and as a pure hemiparesis, without choreic twitchings, even the lucid description of the masters will leave us in doubt unless we also consider the points brought out by later investigators. In my opinion, O. Förster[3] has given the best analysis of the motor disturbances in chorea. He divides them into (1) spontaneous choreic movements, and (2) inco-ordination of voluntary movements. But I believe we can conveniently subdivide the last heading into (*a*) inco-ordination proper, and (*b*) muscular weakness.

1. The spontaneous choreic twitchings are irregular jerks; that is, the elements of rhythm and regularity, such as are found in tremors, are always absent in true chorea. Further, the twitchings mostly occur in single muscles, or in muscle groups that do not act synergetically, as opposed to other forms of muscular movements that preferably affect entire muscle groups which constantly act in unison. The spontaneous movements of chorea never resemble a voluntary movement and seem entirely devoid of purpose.

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[1] British Medical Journal, 1886, p. 636.

[2] Medical News, 1887, p. 406.

[3] Das Wesen der Choreatischen Bewegungsstörung. Volkmann's Sammlung klin. Vorträge, October, 1904.

This distinguishes the disease from tic, in which condition movements, though involuntarily executed, have a superficial resemblance to a purposive act. The choreic twitchings may be likened to the clonic contractions in epilepsy as regards the suddenness of their occurrence. They differ, however, in the fact that there may be only one or two jerks followed by complete relaxation in chorea, while in epilepsy the clonic spasms are an accession of a series of muscular contractions upon muscles already in tonic contraction, which contractions continue for some little time and only gradually merge into relaxation.

2. (*a*) *Disturbances of Co-ordination.* There is a lack of promptness and instability in muscle innervation. The impulse is either delayed or side-tracked into byways instead of reaching the intended muscle paths. Or, if the intended movement is executed there are so many accessory irrelevant movements that an inco-ordinated act is the result instead of the orderly co-ordinated intentional movement of health.

(*b*) *Muscular weakness* results from either a delayed nerve impulse or from dissipation of well-directed energy into channels having an inhibitive effect upon the intended movement.

To summarize this analysis, we have, on the one hand, the involuntary so-called choreic spontaneous movements which depend upon an irritation of single muscles and are dissimilar to voluntary movements. On the other hand, we have disturbances of co-ordination; that is, a disturbance either in the execution of voluntary movements or in the maintenance of static equilibrium. The inco-ordination manifests itself in its complete form as follows: The principal muscles concerned in voluntary movements are promptly innervated, but the impulse is miscarried to a varying degree. In severe cases it is so ineffectual as to cause distinct paresis affecting equally all the muscles of an extremity. In the simple movements of the fingers, tongue, and lips the innervation may be delayed and intermittent.

The impulse for the so-called agonists, the muscles directly concerned in a movement, as opposed to the antagonists, which merely assist the agonists, may radiate into other channels or along the commissural tracts to the opposite side; then we have purposeless associated movements. In like manner the antagonists that support or maintain the agonists are not properly innervated in severe cases of chorea and we have intermittent muscular contractions. In a general way we may state that those impulses that are directly concerned in the volition of station or locomotion are functionating properly; in fact, there is even an excess of function leading to associated movements, while those involuntary motor impulses concerned in the maintenance of equilibration which emanate from the cerebellum and its paths are markedly deficient in chorea. This has led Otfrid Förster to formulate a theory that chorea is primarily a disease of the cerebellum or of the superior cerebellar peduncles. We may assume with him that the toxic agents causing chorea act principally upon the cerebellar tissue and produce both irritant and paralytic effects. The irritant impulses upon the muscles are probably conducted from the cerebellum along the superior cerebellar peduncle, through the red nucleus, to be transmitted either directly into the centrifugal paths, or else to be carried up still higher through intermediate stations into the cerebrum, whence the motor impulse is transmitted in the same way as the twitchings of symptomatic chorea and epilepsy.

Whether this theory proves correct or will shortly be substituted by some other theory, it certainly explains most of the phenomena found in chorea, and particularly does it throw some light upon paralytic chorea.

A study of the various movements in chorea led Weir Mitchell and J. H. Wallace Rhein[1] to distinguish four different types:

1. Movements are continuous during the period of muscular inaction. Hands lying in patient's lap are in constant motion. But when patients perform a delicate muscular effort there is an entire disappearance of movements lasting throughout the muscular effort.

2. In this type there are clonic contractions during rest. Volition so aggravates them that a voluntary movement cannot be accomplished. There is a lessened inhibition during willed muscular effort. This is the usual type.

3. The rarest variety becomes evident when patients attempt to perform a muscular act. The hands at rest move, if at all, only slightly and at rare intervals, but on attempting to use them the twitchings become sufficiently active to prevent or greatly to interfere with the performance of the act.

4. There is a small number of cases in which the movements, continuous during rest, are but slightly altered by the tests employed.

My case showed the third type of movements during the latter part of its course.

CASE.—J. B., aged seven years, a school-boy of Irish-American parentage, was first seen by me January 12, 1906.

Family history is negative: father, mother, brother, sister, uncles, and aunts are free from nervous and mental disorders.

Patient was born in normal labor, walked and talked in proper time, never had convulsions or other nervous manifestations, and mental development showed no anomalies. Of past illnesses, he had diphtheria, followed by mumps, about a year ago, and an intestinal affection, resembling typhoid fever, about nine months ago.

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[1] Philadelphia Medical Journal, 1898, p. 153.

He never had rheumatism or growing pains, nor was there an acute sore throat prior to the development of present complaint.

Present Trouble. About three and a half months ago while on his way from school a companion kicked him in the region of the right ankle. A soft swelling soon appeared over the outer malleolus and the boy began to limp. When he reached home his mother massaged the affected parts and the swelling promptly subsided, but the limping continued. Within the next few days a weakness developed in the entire right lower extremity and a few days later the right upper extremity became similarly involved. All movements with the right arm and hand became clumsy and subsequently impossible.

Four weeks after the beginning of these symptoms he came under my observation.

Examination. The patient, a healthy appearing boy of medium height and weight, is free from signs of visceral disease. His gait, however, is defective, in that he drags his right leg, resembling somewhat the polyneuritic step. The right arm hangs passively by the side and dangles somewhat when the patient walks, so that he gives one the impression of being the subject of a right-sided hemiplegia. The face shows no weakness or paralysis. The pupils are equal and respond promptly to light and accommodation.

Upon testing muscular strength a uniform diminution was found in the flexors and extensors of the upper and the lower extremity on the right side, more of a paresis than a paralysis. The grip in the left hand was good, but that of the right hand was almost nothing, despite the little patient's strenuous efforts to force up the lever of the dynamometer. Neither signs of injury nor atrophies were discoverable anywhere upon repeated examination.

The superficial and deep reflexes appeared normal on both sides, except the right knee-jerk and Achilles tap, which were slightly reduced. In eliciting the patellar reflex I found the upstroke very prompt, while the downward movement was considerably delayed bilaterally, particularly on the paretic side. Upon tapping the tendon the limb flew up promptly, then dallied a while, and came down in a zig-zag manner, continuing this rhythmic movement even after it had regained its former position. There was no spasticity on the affected side and no Babinski, Oppenheim, or Gordon signs. The tongue was protruded in a straight line and showed a few irregular twitchings. No sensory disturbances were found.

About three weeks after my first examination a slight improvement was noticed, but when the patient attempted to use his right hand and arm a peculiar jerkiness and tremor appeared, resembling partly the intention tremor of multiple sclerosis, and having some of the characteristics of tabetic inco-ordination. When directed to touch his nose with the tip of the right index finger there were several purposeless excursions in the arm, forearm, and finger, becoming more exaggerated when he had almost accomplished his design. In an attempt to grip my hand with his right, at first some power was developed, which dwindled down to nothing; then alternating contraction and relaxation of muscles occurred, which could not only be felt but also seen as the tendons intermittently sprang into view. In raising his right lower extremity from the ground inco-ordinate movements developed at once and continued as long as the effort was maintained. In walking he appeared to throw out his right foot instead of using dorsal flexion. The muscular twitchings, previously barely noticeable, had become more numerous. At a subsequent visit the symptom of associated movements could also be demonstrated. When he attempted to make a fist with his left (the unaffected) hand the right participated; when he raised his left leg the right also ascended. But when the movement was initiated by any of the extremities of the right (affected) side no similar participation was indulged in by the left-sided members.

At present his paralytic symptoms are on the decline, so that he walks better and is regaining the use of his right hand. The choreic twitchings, however, have become more marked. This, according to most observers, is a favorable sign.

To summarize: We have a hemiparesis which developed after slight trauma in a previously healthy boy. The weakness began with limping, and within ten days slowly spread over the entire right half of the body with the exception of the face. The reflexes were slightly reduced on the affected side; there were no atrophies, spasticity, or sensory disturbances.

In the differential diagnosis we must consider cerebral hemiplegia, neuritis, and poliomyelitis. Against the diagnosis of cerebral hemiplegia speak the absence of spastic symptoms and the etiology. Anterior poliomyelitis can be ruled out because of the absence of atrophies. The same course can be pursued with neuritis, in which sensory disturbances are a marked feature, while they were never present in this case. We must now consider hysteria and chorea. The patient shows no ancestral or personal tendency to the great neurosis, being of sturdy Irish-American stock, and one out of six young children who are not reared in luxury and receive only the little attention that a workingman's wife can bestow upon them. The boy never showed signs of nervousness and a careful search for hysterical stigmata was negative. Even by exclusion we are forced to make the diagnosis of paralytic chorea. But how can we account for the peculiar onset of the disease with a trauma? As in all forms of trauma, we have the physical and psychical elements to reckon with. The physical element in this case was almost *nil,* a slight transient swelling over the ankle; the psychical element, the unexpected attack—the fright—precipitated the disease, as it often does in other cases of chorea. The physical trauma may have determined the beginning of the condition at the site of its occurrence, but was of secondary importance otherwise.

As regards *treatment*, rest, tonics, and a modified form of Fränkel exercises are the only measures now employed. In the beginning I ordered arsenic in the form of Fowler's solution, which had to be discontinued because it affected the patient unfavorably. Antipyrine in five-grain doses was then substituted, with doubtful results; he then took hypophosphites, with marked benefit. By July, 1906, the patient showed great improvement; he was able to use his right hand, and the gait was very nearly normal. There was only an insignificant amount of inco-ordination in the right upper and lower extremities, and no twitchings could be observed anywhere. The course in my case is rather prolonged (since December, 1905), but, like other prolonged cases, tends toward complete recovery.

In conclusion, I wish to emphasize that associated movements should be looked for in every case of supposed chorea, that some degree of inco-ordination is almost a constant symptom in this disease, and that various degrees of muscle weakness will be found. Muscular twitchings, be they ever so few, constitute a valuable diagnostic aid. A helpful symptom in doubtful cases is the knee-jerk retarded in its descent, first studied and described by Gordon. The slow development of a paralysis in an otherwise healthy child should make us think strongly of paralytic chorea.

POSTURAL TREATMENT OF OTITIS MEDIA AND MASTOIDITIS.

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During the past few years operative procedure has become so common in otology that with many surgeons the slightest sign of involvement of the mastoid antrum complicating an otitis media is considered an indication for operative interference. A better understanding of the anatomical relations of the parts, together with the application of one of the fundamental principles of successful surgery, that is, drainage, would lead to a more conservative line of treatment in a direction that seems to have been entirely overlooked, namely, the posture of a patient with otitis media or mastoiditis. The relationship of the middle ear and antrum is such that, if an inflammatory exudate is present in the middle ear and the patient is lying on his back, the secretions will flow by gravity from the middle ear through the aditus ad antrum to the mastoid antrum itself. The presence of a secretion will, of course, irritate the membrane lining the antrum and produce mastoiditis,